Monostotic Fibrous Dysplasia of the Cervical Spine: Case Report[1]

Charles S. Resnik, M.D.
John R. Lininger, M.D.[2]

Five cases of monostotic fibrous dysplasia involving the spine have previously been reported. We report a sixth case of this entity. The patient had a well-defined lytic lesion with several vertical septations that involved most of the sixth cervical vertebral body with extension to the right pedicle and lamina. Histologic examination revealed fibrous tissue within medullary bone with irregularly shaped bony trabeculae imbedded in the fibrous stroma. Biopsy findings confirmed the diagnosis.

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FIBROUS DYSPLASIA is a term used first in 1938 by Lichtenstein (1) to designate a developmental anomaly of unknown etiology characterized by fibrous tissue replacement of the medullary cavity of bones. Numerous reports of studies of large series of patients with this disorder have been published that included cases of monostotic (one bone) or polyostotic (several bones) involvement (2–10). Although any bone in the body can be involved, vertebral involvement is quite unusual, and only five cases of monostotic fibrous dysplasia of the spine have previously been reported. This report describes a sixth patient who had only one vertebra involved, and who also represents only the third patient ever described who had monostotic fibrous dysplasia of the cervical spine.

CASE REPORT

A 27-year-old woman presented with a complaint of acute neck pain following a minor automobile accident. She had no prior complaints related to the cervical spine or the entire musculoskeletal system. There was no evidence of any endocrine abnormality. Physical examination revealed only minor nuchal rigidity secondary to cervical muscle spasm. No abnormal skin pigmentation was present.

Radiographs of the cervical spine revealed no fracture or other evidence of acute skeletal trauma. A well-defined lytic bone lesion with several septations was seen to involve most of the sixth cervical vertebral body with extension posteriorly to the right pedicle and lamina (Fig. 1). The surrounding cortical margins were completely intact without expansion or destruction. No calcified matrix was demonstrated by conventional tomography (Fig. 2). CT showed no associated soft tissue mass (Fig. 3). Findings on radionuclide bone scans were entirely normal, and a complete radiographic skeletal survey revealed no additional abnormalities.

Hemilaminectomy and biopsy of the sixth cervical vertebra was performed. There was no surgical evidence of gross tumor involvement. Histologic examination revealed a proliferation of fibrous tissue within medullary bone. Imbedded in this fibrous stroma were irregularly shaped bony trabeculae containing osteocytes but lacking peripheral osteoblasts (Fig. 4). When examined under polarized light, the trabeculae consisted of woven bone (Fig. 5). These combined features were diagnostic of fibrous dysplasia.

DISCUSSION

Before 1938, osteitis fibrosa and other similar terms were used to describe the disorder now known as fibrous dysplasia. Since the original description of this entity (1) and the recognition that one or many bones may be involved (6), many authors have discussed fibrous dysplasia and described many patients with the monostotic form (2–5, 7–10). One report (2) included 15 cases of monostotic fibrous dysplasia among 24 patients. Another paper (3) described 55 cases of fibrous dysplasia, 46 of which were monostotic. Other authors have reported 13 patients with monostotic fibrous dysplasia (including one with lumbar vertebral involvement) (4), 50 cases of single bone involvement (5), 16 monostotic lesions among 25 patients (7), 20 patients with fibrous dysplasia (including 16 with the monostotic form) (8), and 22 cases of monostotic involvement (9). The largest series of patients with monostotic fibrous dysplasia was of 67 cases, only one of which involved a cervical vertebra (10).

A total of only five cases of vertebral involvement with monostotic fibrous dysplasia could be found in the medical literature. One patient (4) was a 42-year-old man with an expansile radiolucent lesion in a transverse process of the fourth lumbar vertebra discovered on radiographs obtained because he complained of lower back pain. A 20-year-old man (10) had fibrous dysplasia in the fourth cervical vertebra discovered approximately one year after he sustained a fracture of the fifth cervical vertebra. A 58-year-old woman (11) had the gradual onset (over six years) of symptoms of spinal cord compression. Radiographs revealed a destructive lesion of the first lumbar vertebral body containing fine vertical striations that extended into the pedicle. Radiographs of a 35-year-old woman (12) revealed early destruction of the fourth cervical vertebra approximately one year following neck trauma. This progressed over two months to destruction of a portion of the vertebral body, the superior articular process, the transverse process, and the spinous process with a fine-meshed sclerotic network pattern containing small calcifications. A soft tissue mass was also evident. Another case of monostotic fibrous dysplasia was reported in a 28-year-old woman (13) with a complaint of pain in the lumbar region and disturbance of passing urine for ten years. Radiographs revealed balloon-like dilatation of the transverse and articular processes of the fourth lumbar vertebra.

Most patients with monostotic fibrous dysplasia remain asymptomatic unless a pathological fracture occurs, although, to our knowledge, our patient is the first to be described with asymptomatic monostotic spine involvement. Local trauma may occasionally precede the development of fibrous dysplasia, but a more common occurrence is the incidental discovery of a skeletal lesion on radiographs obtained for acute trauma as in the current case. The usual radiographic features of fibrous dysplasia of the spine are an expansile radiolucent lesion with multiple septations or striations involving the vertebral body and extending into the posterior elements; a soft tissue mass may be present. This appearance resembles that of vertebral hemangioma, and a definite diagnosis can be made only by biopsy. The histologic findings are the same regardless of the site or number of bones involved and consist of a fibrous stroma containing irregularly shaped trabeculae of woven bone that lack peripheral osteoblasts.

[1] From the Departments of Radiology (C.S.R.) and Pathology (J.R.L.), Medical College of Virginia Hospitals, Richmond, Virginia. Received July 1, 1983; accepted Aug. 19. cp

[2] Present address: Department of Pathology, Montgomery Hospital, Norristown, Pennsylvania 19401.

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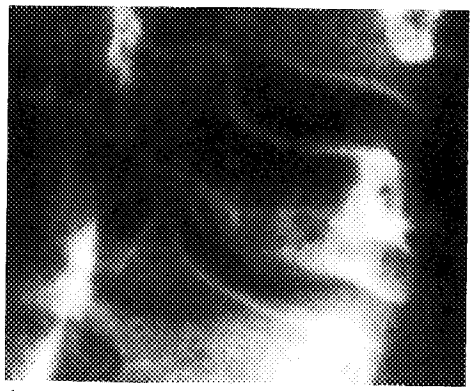
Figures 1-5



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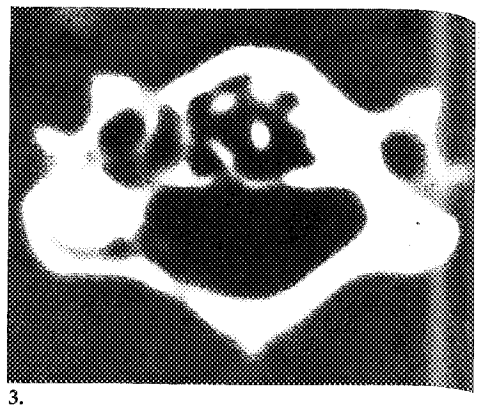

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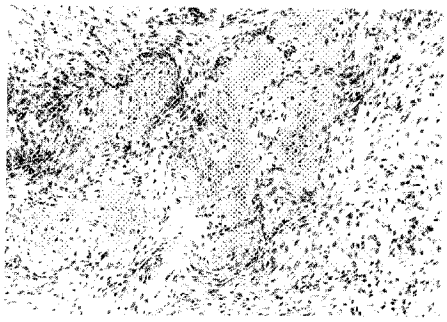
1. Oblique radiograph of the cervical spine showing a well-defined lytic lesion with several septations involving most of the sixth cervical vertebral body with extension to the right pedicle and lamina.
2. Anteroposterior tomogram showing the thin vertical septations and no calcified matrix.
3. Computed tomogram showing vertebral body and pedicle involvement with no associated soft tissue mass.
4. Hematoxylin and eosin section (140×) showing fibrous stroma containing irregularly shaped bone trabeculae that lack peripheral osteoblasts.
5. Examination under polarized light (350×) showing woven bone characteristic of fibrous dysplasia.



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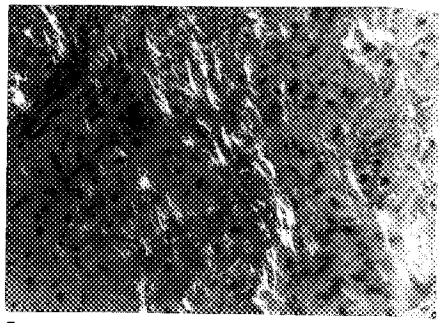

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Charles S. Resnik, M.D.
Department of Radiology
Medical College of Virginia Hospitals
Box 615, MCV Station
Richmond, Virginia 23298

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